

MUSCLE & NERVE



CONTENTS

Official Journal of the American Association of Electrodiagnostic Medicine

JULY 2003

- | | | |
|--------------------------------|------------|--|
| INVITED REVIEWS | 1 | Multichannel Surface EMG: Basic Aspects and Clinical Utility
<i>Machiel J. Zwarts, MD, PhD, and Dick F. Stegeman, PhD</i> |
| | 18 | Sphincter Electromyography and Multiple System Atrophy
<i>Frederick Nahm, MD, PhD, and Roy Freeman, MD</i> |
| SPECIAL INVITED ARTICLE | 27 | Ethical Review of Research Involving Human Subjects: When and Why Is IRB Review Necessary?
<i>Richard M. Wagner, MA</i> |
| MAIN ARTICLES | 40 | Ulnar Neuropathy in the Forearm: A Possible Complication of Diabetes Mellitus
<i>Juan A. Acosta, MD, Stuart N. Hoffman, DO, Elizabeth M. Raynor, MD, Rachel A. Nardin, MD, and Seward B. Rutkove, MD</i> |
| | 46 | Time and Magnitude of Torque Generation Is Impaired in Both Arms following Stroke
<i>Patrick H. McCrea, BSc, Janice J. Eng, PhD, and Antony J. Hodgson, PhD</i> |
| | 54 | Knee Extensor Torque, Work, and EMG during Subjectively Graded Dynamic Contractions
<i>Danny M. Pincivero, PhD, Paul T. Dixon, and Alan J. Coelho, EdD</i> |
| | 62 | Muscle Fiber Number in the Biceps Brachii Muscle of Young and Old Men
<i>Cliff S. Klein, PhD, Greg D. Marsh, PhD, Robert J. Petrella, MD, PhD, and Charles L. Rice, PhD</i> |
| | 69 | Compressive Lumbar Myelopathy Presenting as Segmental Motor Neuron Disease
<i>Kleopas A. Kleopa, MD, Eleni Zamba-Papanicolaou, MD, and Theodoros Kyriakides, MRCP</i> |
| | 74 | Strength Training Alters the Viscoelastic Properties of Tendons in Elderly Humans
<i>Neil D. Reeves, MSc, Marco V. Narici, PhD, and Constantinos N. Maganaris, PhD</i> |
| | 82 | F-Wave Acquisition Using Low-Current Stimulation
<i>Margarete DiBenedetto, MD, Shawn D. Gale, PhD, Demitri Adarmes, MD, and Michael Schabacker, MD</i> |
| | 87 | Effects of Endurance Training on Satellite Cell Frequency in Skeletal Muscle of Old Men
<i>Nadia Charifi, MSc, Fawzi Kadi, PhD, Léonard Féasson, MD, MSc, and Christian Denis, MD, PhD</i> |
| | 93 | Comparison between Impairment and Disability Scales in Immune-Mediated Polyneuropathies
<i>Ingemar S.J. Merkies, MD, PhD, Paul I.M. Schmitz, PhD, Frans G.A. Van Der Meché, MD, PhD, and Pieter A. Van Doorn, MD, PhD</i> |
| | 101 | Quantitative Measurement of Muscle Fiber Composition in a Normal Population
<i>Ingrid Toft, MD, PhD, Sigurd Lindal, MD, PhD, Kaare H. Bønnå, MD, PhD, and Trond Jenssen, MD, PhD</i> |
| SHORT REPORTS | 109 | Hexosaminidase A Deficiency Is an Uncommon Cause of a Syndrome Mimicking Amyotrophic Lateral Sclerosis
<i>Vivian E. Drory, MD, Miriam Birnbaum, PhD, Leah Peleg, PhD, Boleslaw Goldman, MD, and Amos D. Korczyn, MD, MSc</i> |

CASES OF THE MONTH

- 113 **Novel Missense Mutation and Large Deletion of GNE Gene in Autosomal-Recessive Inclusion-Body Myopathy**
Roberto Del Bo, PhD, Pierluigi Baron, MD, Alessandro Prella, MD, Massimo Serafini, BS, Maurizio Moggio, MD, Alessio Di Fonzo, MD, Marina Castagni, PhD, Nereo Bresolin, MD, and Giacomo Pietro Comi, MD

- 118 **Adult Hallervorden-Spatz Syndrome Simulating Amyotrophic Lateral Sclerosis**
O.M. Vasconcelos, MD, D.H. Harter, MD, C. Duffy, BA, B. McDonough, RN, J.G. Seidman, PhD, C.E. Seidman, MD, and W.W. Campbell, MD, MSHA

- 123 **Masticator Myopathy**
Shigeo Yamada, MD, Tomoko Ogawa, Jin Nishimiya, MD, PhD, Tatsuhiko Yuasa, MD, PhD, and Fumitoshi Taketazu, MD, PhD

REGULAR FEATURES

- 128 **Letter to the Editor**
 130 **Calendar of Events**
 131 **AAEM News and Comments**

AUGUST 2003

INVITED REVIEWS

- 133 **Lyme Disease and the Peripheral Nervous System**
John J. Halperin, MD
 144 **The H-Reflex as a Tool in Neurophysiology: Its Limitations and Uses in Understanding Nervous System Function**
John E. Misiaszek, PhD

MAIN ARTICLES

- 161 **Motor Unit Number Estimate of Distal and Proximal Muscles in Charcot-Marie-Tooth Disease**
Richard A. Lewis, MD, Jun Li, MD, PhD, Darren R. Fuerst, PhD, Michael E. Shy, MD, and Karen Krajewski, MS
 168 **Effects of Imagery Motor Training on Torque Production of Ankle Plantar Flexor Muscles**
Inge Zijdwind, PhD, Sjoukje T. Toering, MSc, Bram Bessert, MD, Occo van der Laan, MD, and Ron L. Diercks, PhD, MD
 174 **Changing Stimulation Patterns Improves Performance during Electrically Elicited Contractions**
Wayne B. Scott, MPT, and Stuart A. Binder-Macleod, PhD
 181 **Changes in Neuromuscular Function after Training by Functional Electrical Stimulation**
Tanguy Marqueste, PhD, François Hug, PhD, Patrick Decherchi, PhD, and Yves Jammes, MD, DSc
 189 **Dispersion of the Distal Compound Muscle Action Potential in Chronic Inflammatory Demyelinating Polyneuropathy and Carpal Tunnel Syndrome**
James C. Cleland, MB, ChB, Eric L. Logigian, MD, Pariwat Thaisethawatkul, MD, and David N. Herrmann, MB, BCh
 194 **Evaluation of Morphological and Functional Regeneration of Rat Nerve-Muscle Units after Temporary and Permanent Tubulization**
Mitsuaki Odaka, MD, Yoshiyasu Uchiyama, MD, PhD, Yoshinori Oka, MD, PhD, and Tetsuro Tamaki, PhD
 204 **Decomposition-Based Quantitative Electromyography: Methods and Initial Normative Data in Five Muscles**
Timothy J. Doherty, MD, PhD, and Daniel W. Stashuk, PhD
 212 **Noninactivating, Tetrodotoxin-Sensitive Na⁺ Conductance in Peripheral Axons**
Hajime A. Tokuno, MD, Jeffery D. Kocsis, PhD, and Stephen G. Waxman, MD, PhD
 218 **Quadriceps Muscle Strength and Voluntary Activation after Polio**
Anita Beelen, PhD, Frans Nollet, MD, PhD, Marianne de Visser, MD, PhD, Bareld A. de Jong, MD, PhD, Gustaaf J. Lankhorst, MD, PhD, and Anthony J. Sargeant, PhD

	227	Preferential Motor Reinnervation in the Mouse: Comparison of Femoral Nerve Repair Using a Fibrin Sealant or Suture <i>Grant A. Robinson, PhD, and Roger D. Madison, PhD</i>
SHORT REPORTS	232	Paramyotonia Congenita due to a De Novo Mutation: A Case Report <i>Takayasu Fukudome, MD, Hajime Izumoto, MD, Hirofumi Goto, MD, Hidenori Matsuo, MD, Toshiro Yoshimura, MD, Shun-Ichi Sakoda, MD, and Noritoshi Shibuya, MD</i>
	236	Decremental Response of the Nasalis and Hypothenar Muscles in Myasthenia Gravis <i>Erik H. Niks, MD, Umesh A. Badrising, MD, Jan J. Verschuuren, MD, PhD, and J. Gert van Dijk, MD, PhD</i>
	239	Monophasic and Biphasic Stimulation Evoke Different Responses <i>Edelle C. Field-Fote, PhD, Brent Anderson, BS, Valma J. Robertson, PhD, and Neil I. Spielholz, PhD</i>
CASES OF THE MONTH	242	Progressive Arm Weakness and Tonic Hand Spasm from Multifocal Motor Neuropathy in the Brachial Plexus <i>R. Veltkamp, MD, M. Krause, MD, C. Schranz, MD, and H.-M. Meinck, MD</i>
	246	Axonal Pharyngeal-Cervical-Brachial Variant of Guillain-Barré Syndrome without Anti-Gt1a IgG Antibody <i>Motomi Arai, MD, PhD, Keiichi Susuki, MD, PhD, and Michiaki Koga, MD, PhD</i>
REGULAR FEATURES	251	Letters to the Editor
	254	Book Review
	255	Calendar of Events
	256	AAEM News and Comments

SEPTEMBER 2003

OBITUARY	261	Bernard Katz <i>A.R. Martin, PhD</i>
INVITED REVIEWS	263	Statistical Motor Unit Number Estimation: From Theory to Practice <i>Catherine Lomen-Hoerth, MD, PhD, and Michael P. Slawnych, PhD</i>
	273	Immunotherapy of Idiopathic Inflammatory Neuropathies <i>Peter D. Donofrio, MD</i>
MAIN ARTICLES	293	Rapsyn Mutations in Myasthenic Syndrome due to Impaired Receptor Clustering <i>Ricardo A. Maselli, MD, Vanessa Dunne, BS, Samuel Ignacio Pascual-Pascual, MD, Constance Bowe, MD, Mark Agius, MD, Rochelle Frank, MD, and Robert L. Wollmann, MD, PhD</i>
	302	Clinical Spectrum of Muscle Weakness in Human West Nile Virus Infection <i>A. Arturo Leis, MD, Dobrivoje S. Stokic, MD, Risa M. Webb, MD, Sally A. Slavinski, DVM, MPH, and Jonathan Fratkin, MD</i>
	309	Relative Contributions of Neural Mechanisms versus Muscle Mechanics in Promoting Finger Extension Deficits following Stroke <i>D.G. Kamper, PhD, R.L. Harvey, MD, S. Suresh, MD, and W.Z. Rymer, MD, PhD</i>
	319	A Prospective Evaluation of Phrenic Nerve Conduction in Multifocal Motor Neuropathy and Chronic Inflammatory Demyelinating Polyneuropathy <i>Frederic Macia, MD, Gwendal Le Masson, MD, Marie Rouanet-Larriviere, MD, Desire Habonimana, MD, Xavier Ferrer, MD, Roger Marthan, MD, and Alain Lagueny, MD</i>
	324	Cyclosporin A Treatment Increases Rat Soleus Muscle Oxidative Capacities <i>Hervé Sanchez, PhD, Benoit N'Guessan, BS, Florence Ribera, PhD, Renée Ventura-Clapier, PhD, and Xavier Bigard, MD, PhD</i>
	330	Peripheral Neuropathy in Scleroderma <i>Ann N. Poncelet, MD, and M. Kari Connolly, MD</i>
	336	Ventilatory Dysfunction in mdx Mice: Impact of Tumor Necrosis Factor-Alpha Deletion <i>Luc E. Gosselin, PhD, Jacob E. Barkley, MS, Melissa J. Spencer, PhD, Kathleen M. McCormick, PhD, and Gaspar A. Farkas, PhD</i>

	344	Neurophysiological Changes in Demyelinating and Axonal Forms of Acute Experimental Autoimmune Neuritis in the Lewis Rat <i>Jude M. Taylor, PhD, and John D. Pollard, MD, PhD</i>
	353	Caffeine Impairs Intramuscular Energy Balance in Patients Susceptible to Malignant Hyperthermia <i>Zoran Textor, MD, Meinrad Beer, MD, Martin Anetseder, MD, Herbert Köstler, PhD, Erwin Kagerbauer, MS, Werner Kenn, MD, Dietbert Hahn, MD, and Norbert Roewer, MD</i>
	359	Caffeine Increases Spinal Excitability in Humans <i>C. Walton, MSc, J. Kalmar, MSc, and E. Cafarelli, PhD</i>
	365	Improvement of Muscle Healing through Enhancement of Muscle Regeneration and Prevention of Fibrosis <i>Kenji Sato, MD, Yong Li, MD, PhD, William Foster, BS, Kazumasa Fukushima, MD, Neil Badlani, MS, Nobuo Adachi, MD, Arvydas Usas, MD, Freddie H. Fu, MD, and Johnny Huard, PhD</i>
SHORT REPORTS	373	Inflammatory Demyelination in a Patient with CMT1A <i>Anne Vital, MD, PhD, Claude Vital, MD, Alain Lagueny, MD, Xavier Ferrer, MD, Catherine Ribière-Bachelier, MD, Philippe Latour, MD, and Klaus G. Petry, PhD</i>
	377	Electromyography of the Anal Sphincter: Which Muscle to Examine? <i>Simon Podnar, MD, DSc</i>
	380	Two Novel Mutations in the Muscle Glycogen Phosphorylase Gene in McArdle's Disease <i>Josep Gámez, MD, PhD, Juan C. Rubio, MSc, Miguel A. Martín, PhD, Israel Fernández-Cadenas, BSc, Elena García-Arumi, PhD, Antoni L. Andreu, MD, PhD, and Joaquín Arenas, PhD</i>
CASE OF THE MONTH	383	Spinal Accessory Neuropathy, Droopy Shoulder, and Thoracic Outlet Syndrome <i>Amer Al-Shekhlee, MD, and Bashir Kadirji, MD</i>
REGULAR FEATURES	386	Letters to the Editor
	389	Book Review
	390	Calendar of Events

OCTOBER 2003

INVITED REVIEW	391	Cutaneous Silent Periods <i>Mary Kay Floeter, MD, PhD</i>
MEDICAL HISTORY	402	Origins and Early Descriptions of "Duchenne Muscular Dystrophy" <i>Kenneth L. Tyler, MD</i>
MAIN ARTICLES	423	Increased Jitter and Blocking in Normal Muscles due to Doubly Innervated Muscle Fibers <i>Zoia C. Lateva, PhD, Kevin C. McGill, PhD, and M. Elise Johanson, MS</i>
	432	Riluzole Attenuates Spinal Muscular Atrophy Disease Progression in a Mouse Model <i>Hafedh Haddad, MD, Carmen Cifuentes-Díaz, PhD, Audrey Miroglio, MSc, Natacha Roblot, Vandana Joshi, MSc, and Judith Melki, MD, PhD</i>
	438	Contribution of Wild-Type Transthyretin to Hereditary Peripheral Nerve Amyloid <i>Masahide Yazaki, MD, PhD, Juris J. Liepnieks, PhD, John C. Kincaid, MD, and Merrill D. Benson, MD</i>
	443	Late-Onset Distal Muscular Dystrophy Affecting the Posterior Calves <i>Jonathan S. Katz, MD, Thomas A. Rando, MD, PhD, Richard J. Barohn, MD, David S. Saperstein, MD, Carlayne E. Jackson, MD, Matthew Wicklund, MD, and Anthony A. Amato, MD</i>
	449	Neutrophils Do Not Contribute to Local Tissue Damage, but Play a Key Role in Skeletal Muscle Regeneration, in Mice Injected with <i>Bothrops Asper</i> Snake Venom <i>C. F. P. Teixeira, PhD, S. R. Zamunér, PhD, J. P. Zuliani, MSc, C. M. Fernandes, MSc, M. A. Cruz-Hofling, PhD, I. Fernandes, PhD, F. Chaves, MSc, and J. M. Gutiérrez, PhD</i>

- 460 **Near-Nerve Potential in Lepromatous Leprosy**
Wilson Marques, Jr., MD, Norma T. Foss, MD, Ana P. M. Arruda, MD,
and Amilton A. Barreira, MD
- 464 **Inferior Mechanical Properties of Spastic Muscle Bundles due to Hypertrophic
but Compromised Extracellular Matrix Material**
Richard L. Lieber, PhD, Eva Runesson, PhD, Fredrik Einarsson, MD, Jan Fridén, MD, PhD
- 472 **Calpain 3 Cleaves Filamin C and Regulates Its Ability to Interact with γ - and δ -
Sarcoglycans**
Jeffrey R. Guyon, PhD, Elena Kudryashova, MS, Alexandra Potts, Isin Dalkilic, BA,
Melissa A. Brosius, MS, Terri G. Thompson, PhD, Jacques S. Beckmann, PhD,
Louis M. Kunkel, PhD, and Melissa J. Spencer, PhD
- 484 **Overexpressions of Myoglobin and Antioxidant Enzymes in Ragged-Red Fibers
of Skeletal Muscle from Patients with Mitochondrial Encephalomyopathy**
Makoto Kunishige, MD, Takao Mitsui, MD, Masashi Akaike, MD, Masakazu Kawajiri, MD,
Masayuki Shono, PhD, Hisaomi Kawai, MD, and Toshio Matsumoto, MD
- 493 **Mechanisms of Force Loss in Diabetic Mouse Skeletal Muscle**
Lisa A. Lesniewski, MA, Todd A. Miller, PhD, and R. B. Armstrong, PhD

SHORT REPORTS

- 501 **Stimulated Single-Fiber EMG of the Frontalis and Orbicularis Oculi Muscles in
Ocular Myasthenia Gravis**
J. Valls-Canals, MD, M. Povedano, MD, J. Montero, MD, PhD, and J. Pradas MD, PhD
- 504 **Clinical Significance of Complex Repetitive Discharges: A Case-Control Study**
Lesley K. Fellows, MD, CM, DPhil, Bethany J. Foster, MD,
and Colin H. Chalk, MD, CM, FRCPC
- 508 **Progressive Exercise Intolerance Associated with a New Muscle-Restricted
Nonsense Mutation (G142X) in the Mitochondrial Cytochrome *b* Gene**
Claudio Bruno, MD, Filippo M. Santorelli, MD, Stefania Assereto, BS,
Emmanuel Tonoli, MS, Alessandra Tessa, PhD, Monica Traverso, BS, Sara Scapolan, MD,
Massimo Bado, MD, Silvana Tedeschi, PhD, and Carlo Minetti, MD

CASE OF THE MONTH

- 512 **Retrograde Regeneration following Neurotmesis of the Ulnar Nerve**
A. Arturo Leis, MD, John A. Lancon, MD, Dobrivoje S. Stokic, MD

REGULAR FEATURES

- 515 **Letters to the Editor**
- 518 **Book Review**
- 519 **Calendar of Events**
- 520 **AAEM News and Comments**

NOVEMBER 2003

OBITUARY

- 523 **Edward H. Lambert**
Jasper R. Daube, MD

INVITED REVIEWS

- 525 **Peripheral Nerve Fascicles: Anatomy and Clinical Relevance**
John D. Stewart, MB, BS, FRCP(C)
- 542 **The Peripheral Nerve Complications of Human Immunodeficiency Virus (HIV)
Infection**
B. J. Brew, MD, FRACP

MAIN ARTICLES

- 553 **Positive Salivary Gland Biopsy, Sjögren Syndrome, and Neuropathy: Clinical
Implications**
Kenneth C. Gorson, MD, and Allan H. Ropper, MD
- 561 **Acetylcholine Receptor Distribution and Synapse Elimination at the Developing
Neuromuscular Junction of *mdx* Mice**
Elaine Minatel, PhD, Humberto Santo Neto, PhD, and Maria Julia Marques, PhD
- 570 **Clinical Study of FK506 in Patients with Myasthenia Gravis**
Tetsuro Konishi, MD, Yasumasa Yoshiyama, MD, Masaharu Takamori, MD,
Koichi Yagi, MD, Eiichiro Mukai, MD, Takahiko Saïda, MD,
and The Japanese FK506 MG Study Group

	575	Melas with Point Mutations Involving tRNA^{Leu} (A3243G) and tRNA^{Glu} (A14693G) <i>Chin-Yuan Tzen, MD, PhD, Peterus Thajeb, MD, Tsu-Yen Wu, BSc, and Shiu-Ching Chen, BSc</i>
	582	Vasomotor Dysfunction in Carpal Tunnel Syndrome <i>Einar P.V. Wilder-Smith, MD, Stephanie Fook-Chong, BSc, Soh Eng Chew, BSc, Adeline Chow, BSc, and Yaling Guo, MD</i>
	587	Differential Responses of HSPs to Heat Stress in Slow and Fast Regions of Rat Gastrocnemius Muscle <i>Yasuharu Oishi, PhD, Kouhachi Taniguchi, PhD, Hisahiro Matsumoto, PhD, Akihiko Ishihara, PhD, Yoshinobu Ohira, PhD, and Roland R. Roy, PhD</i>
	595	Autonomic and Peripheral Neuropathy in Endstage Liver Disease and following Liver Transplantation <i>Alan J. McDougall, PhD, Leo Davies, MD, and Geoff W. McCaughan, PhD</i>
	601	Strength Training in Older Women: Early and Late Changes in Whole Muscle and Single Cells <i>Walter R. Frontera, MD, PhD, Virginia A. Hughes, MS, Lisa S. Krivickas, MD, Sang-Kyu Kim, MD, PhD, Mona Foldvari, MS, and Ronenn Roubenoff, MD</i>
	609	Acute Leg-Cycling Exercise Attenuates the H-Reflex Recorded in Soleus but not Flexor Carpi Radialis <i>Robert W. Motl, PhD, and Rod K. Dishman, PhD</i>
	614	Role of Intertendinous Connections in Distribution of Force in the Human Extensor Digitorum Muscle <i>Douglas A. Keen, PhD, and Andrew J. Fuglevand, PhD</i>
SHORT REPORTS	623	X-Linked Motor and Sensory Neuropathy with Pyramidal Signs and Cerebral White Matter Lesions <i>Hirofumi Goto, MD, Hidenori Matsuo, MD, Akio Ohnishi, MD, Takayasu Fukudome, MD, and Noritoshi Shibuya, MD</i>
	626	Occurrence of A-Waves in F-Wave Studies of Healthy Nerves <i>Leena Puksa, MD, Erik Stålberg, MD, PhD, and Björn Falck, MD, PhD</i>
	630	Clinical and Neurophysiological Evaluation of Progression in Amyotrophic Lateral Sclerosis <i>Mamede de Carvalho, MD, Manuel Scotto, PhD, Arminda Lopes, MD, and Michael Swash, MD</i>
CASES OF THE MONTH	634	Mononeuropathy Multiplex in Association with Livedoid Vasculitis <i>Cory Toth, MD, Martin Trotter, PhD, MD, Arthur Clark, MD, and Douglas Zochodne, MD</i>
	640	McArdle's Disease Presenting as Recurrent Cryptogenic Renal Failure due to Occult Seizures <i>Adrianne R. Walker, BA, Kevin Tschetter, MD, Fumitsu Matsuo, MD, and Kevin M. Flanigan, MD</i>
ISSUES & OPINIONS	644	Piriformis Syndrome <i>John D. Stewart, MB, BS, FRCP(C), Loren M. Fishman, MD, and Michael P. Schaefer, MD</i>
REGULAR FEATURES	650	Letters to the Editor
	653	Calendar of Events
	654	AAEM News and Comments

DECEMBER 2003

EDITORIAL	657	Who Is the Primary Suspect in Acute Motor Axonal Neuropathy? <i>Ryuji Kaji, MD, PhD, and Hiroyuki Nodera, MD</i>
INVITED REVIEW	659	Cytokines, Chemokines, and Cell Adhesion Molecules in Inflammatory Myopathies <i>Dominique Figarella-Branger, MD, PhD, Muriel Civatte, MD, Catherine Bartoli, PhD, and Jean-François Pellissier, MD</i>

MAIN ARTICLES

- 683 **The Refractory Period of Transmission Is Impaired in Axonal Guillain-Barré Syndrome**
Satoshi Kuwabara, MD, Hugh Bostock, PhD, Kazuo Ogawara, MD, Jia-Ying Sung, MD, Kazuaki Kanai, MD, Masahiro Mori, MD, Takamichi Hattori, MD, and David Burke, MD, DSc
- 690 **Prevalence and Progression of Mitochondrial Diseases: A Study of 50 Patients**
Javier Arpa, MD, Antonio Cruz-Martínez, MD, Yolanda Campos, MSc, Manuel Gutiérrez-Molina, MD, Francisco García-Río, MD, Concepción Pérez-Conde, MD, Miguel A. Martín, PhD, Juan C. Rubio, MSc, Pilar Del Hoyo, PharmB, Ana Arpa-Fernández, MD, and Joaquín Arenas, PhD
- 696 **Atypical Presentations of Primary Amyloid Neuropathy**
Steve Vucic, MB, BS, Peter Siao Tick Chong, MD, and Didier Cros, MD
- 703 **Enzyme Replacement Therapy Improves Peripheral Nerve and Sweat Function in Fabry Disease**
Raphael Schiffmann, MD, Mary Kay Floeter, MD, PhD, James M. Dambrosia, PhD, Surya Gupta, MD, David F. Moore, MD, PhD, Yehonatan Sharabi, MD, Ramesh K. Khurana, MD, and Roscoe O. Brady, MD
- 711 **Preserved Cutaneous Silent Periods in Severe Entrapment Neuropathies**
Markus Kofler, MD, Katrin Fröhlich, MD, and Leopold Saltuari, MD
- 715 **Gelsolin-Related Familial Amyloidosis, Finnish Type, in a Portuguese Family: Clinical and Neurophysiological Studies**
Isabel Conceição, MD, Maria Lurdes Sales-Luis, MD, Mamede De Carvalho, MD, Teresinha Evangelista, MD, Rui Fernandes, PhD, Tiina Paunio, MD, Hannele Kangas, MD, Paula Coutinho, MD, Carlos Neves, MD, and Maria João Saraiva, PhD
- 722 **Characterization of Two New Dominant CIC-1 Channel Mutations Associated with Myotonia**
Morten Grønnet, PhD, Thomas Jespersen, PhD, Eskild Colding-Jørgensen, MD, Marianne Schwartz, PhD, Dan A. Klaerke, MD, John Vissing, MD, PhD, Søren-Peter Olesen, MD, PhD, and Morten Dunø, PhD
- 733 **Acute Neuropathies after Peripheral Blood Stem Cell and Bone Marrow Transplantation**
Alejandro A. Rabinstein, MD, Angela Dispenzieri, MD, Ivana N. Micallef, MD, David J. Inwards, MD, Mark R. Litzow, MD, and Eelco F. M. Wijdicks, MD
- 737 **Deleted 4977-bp Mitochondrial DNA Mutation Is Associated with Sporadic Amyotrophic Lateral Sclerosis: A Hospital-Based Case-Control Study**
Long-Sun Ro, MD, PhD, Shiao-Lin Lai, MD, Chiung-Mei Chen, MD, PhD, and Sien-Tsong Chen, MD
- 744 **Endurance Training Improves Skeletal Muscle Electrical Activity in Active COPD Patients**
Nadège Gosselin, PhD, Karen Lambert, PhD, Magali Poulain, PhD, Anne Martin, Christian Préfaut, MD, and Alain Varray, PhD

SHORT REPORTS

- 754 **Lumbosacral Radiculopathy following Radiofrequency Ablation Therapy**
D. Jim Coskun, MD, James Gilchrist, MD, and Damian Dupuy, MD
- 757 **Late-Onset Mitochondrial Disorder with Electromyographic Evidence of Myotonia**
Mathew L.P. Howse, MRCP, Theresa M. Wardell, BSc, Christopher J. Fisher, FRCP, Peter J.B. Tilley, FRCP, Patrick F. Chinnery, PhD, MRCP, and Laurence Bindoff, MD, MRCP

CASES OF THE MONTH

- 760 **Isolated Compression of the Pectoral Nerve Resulting in Atrophy of the Major Pectoral Muscle**
Alexander Gardetto, MD, Claudia Thaler, MD, Stefan Kiechl, MD, Herbert Maurer, MD, and Hildegunde Piza-Katzer, MD
- 764 **Ocular Myasthenia Gravis Associated with Euthyroid Ophthalmopathy**
Tomohiko Kusuvara, MD, Masashi Nakajima, MD, and Akiko Imamura, MD

AAEM PRACTICE TOPIC

- 767 **Laryngeal Electromyography: An Evidence-Based Review**
AAEM Laryngeal EMG Task Force

**REGULAR
FEATURES**

773	Letters to the Editor
776	Book Review
777	Calendar of Events
778	AAEM News and Comments
787	Volume 28 Author Index
793	Volume 28 Subject Index
I	Volume 28 Contents

